

Your diagnosis? Umbilical cord tumor

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Clinical information

A 35-year-old woman, gravida 3, para 3, at 33 1/7 weeks of gestation was referred to the obstetric clinic for prenatal assessment of an echogenic mass of the umbilical cord detected during a routine ultrasound examination. Chorionic villus biopsy for advanced maternal age at 13-week gestation revealed a normal 46 XX karyotype. Sonographic assessment of the fetus demonstrated growth parameters appropriate for the gestational age. The abdominal wall was

closed, with an inhomogeneous mass measuring 43×32×28 mm near the insertion of the umbilical cord (Fig. 1). The cardiotocogram showed no fetal distress. Doppler flow studies of the umbilical cord showed three vessels, displaced by a multilobulated echogenic tumor. Maternal serum screening showed an elevated level of alpha-fetoprotein (326 µg/l), but no signs of congenital infection.

At 38 5/7 weeks of gestation, a girl was delivered vaginally, weighing 2,840 g (10th–25th centile), with a length of 47 cm (10th–25th centile) and head circumference of 33 cm (10th–25th centile). She adapted with Apgar scores of 8/9/9, and the arterial cord pH was 7.31. The umbilical cord showed a nodular bulge proximal to its insertion into the abdominal wall. It measured 4 cm in diameter, had a smooth, translucent, pearly colored surface, without superficial vessels or pulsations, and was of firm, elastic consistency. A blue core was discernible within (Fig. 2). The findings were associated with marked thickening and edema of the Wharton's jelly, and one vein and two arteries were present within the cord. A 1-cm segment of normal umbilical cord separated the tumor from the intact abdominal wall. Abdominal ultrasound examination showed no visceral abnormalities. The umbilical cord was clamped distal to the lesion, and the infant recovered in a puerperal care unit. The macro- and microscopic placenta examination (387 g, 19×15×3 cm) revealed no abnormalities. At 2 days of age, the infant was transferred to a pediatric surgery unit for surgical revision.

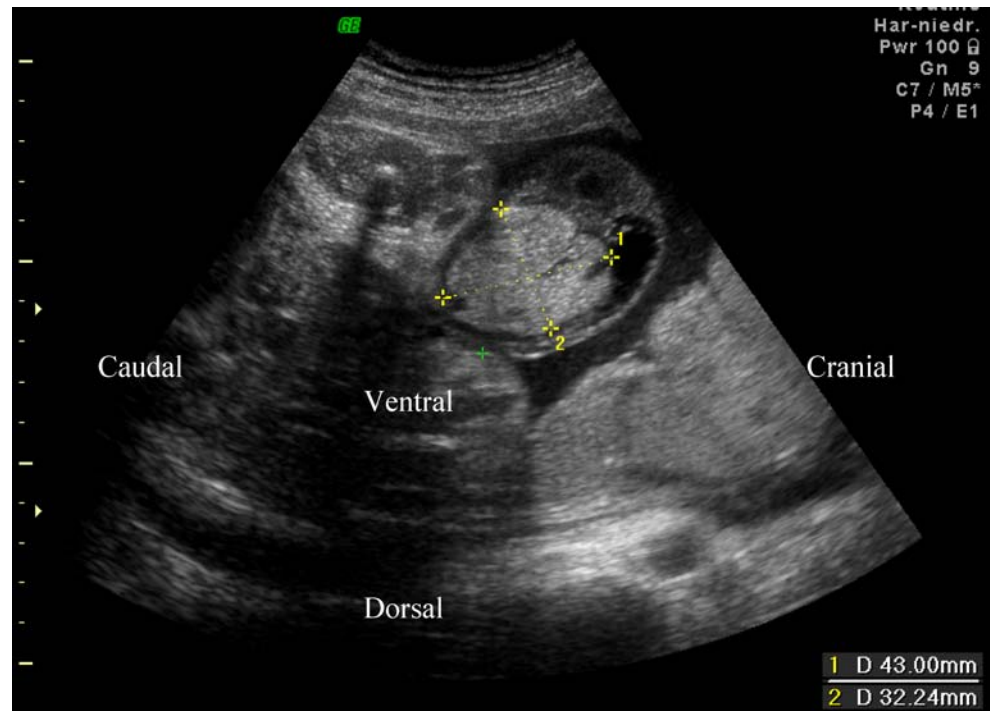
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Fig. 1 Fetal abdominal wall with umbilicus at 33+1 weeks of gestation. The arrow marks the umbilical vein passing through the abdominal wall. The dimensions of the umbilical cord tumor are shown



What is your diagnosis?

Diagnosis: umbilical cord hemangioma

Surgical revision was undertaken for the suspected diagnosis of a hernia into the cord. However, no defect of the abdominal wall was found. Surgical section of the umbilical cord was performed with an umbilicoplasty. Postoperative recovery was uneventful. Concerning the umbilical cord pathology, the maximum diameter of the surgically removed 8.5-cm long umbilical cord segment was 3.6 cm. Macroscopically, the cross section revealed three vessels. Only on microscopic examination did it become clear that

the Wharton's jelly was mostly replaced by abundant aggregates of thin-walled capillaries. The capillary hemangioma showed a diffuse growth around the vessels, with focal dissection of the muscular coat of the umbilical vein. In addition, nodular aggregates were seen, some freshly thrombosed (Fig. 3). Distal to the removed lesion, the remaining umbilical cord was unremarkable.



Fig. 2 Appearance of the umbilical cord shortly after birth. Note the nodular bulging and the distal edema

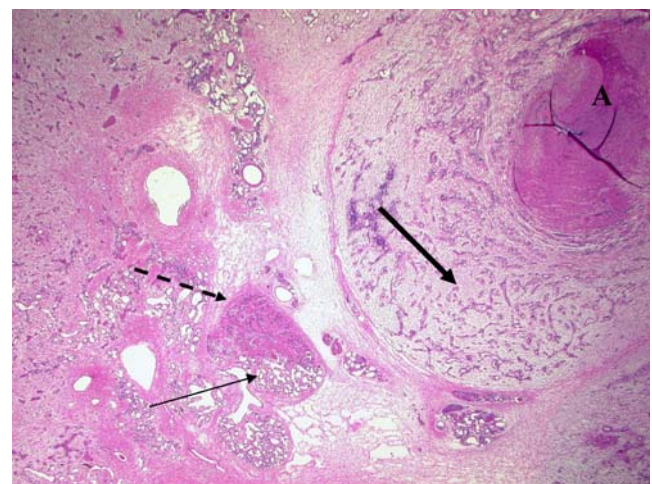


Fig. 3 Cross-section of umbilical cord showing an umbilical artery (a). Note the abundance of tiny capillaries around the artery (thick arrow). Some capillary growth is nodular (thin arrow) with occasional fresh thromboses (dotted arrow). Most of the Wharton's jelly is replaced by capillaries. Hematoxylin-eosin, magnification $\times 100$

Table 1 Differential diagnosis of an umbilical cord tumor

Diagnosis	Macroscopic aspect	Association with other findings
1) Umbilical cord teratoma	Nodular or cystic; maximum 10 cm Ø; normal umbilical cord origin; all along the umbilical cord	• 50% associated with other malformations
2) Hemangioma of the umbilical cord	Nodular 0.2 to 18 cm Ø; normal umbilical cord origin; proximal to the placenta	• Mostly isolated finding; • edema of Wharton's jelly; • complication: intrauterine bleeding
3.1) Hernia into the cord	Nodular <4 cm Ø; normal umbilical cord origin; proximal to the abdomen; elastic consistence	• Mostly isolated finding; • associated with Meckel's diverticulum
3.2) Omphalocele	Abdominal organ prolapse >4 cm Ø; large umbilical cord origin	• Associated with other malformations, syndromes or chromosomal abnormalities
4) Umbilical cord cyst	Nodular or cystic; most often single; normal umbilical cord origin; all along the umbilical cord	• Mostly isolated finding
5) Umbilical cord polyp	Nodular, solid, reddish, exudative, sometimes ulcerated and bleeding	• Meckel's diverticulum and umbilical enteric fistula
6) Umbilical cord hematoma ¹ and aneurysm ²	¹ Nodular, livid; ² cystic in US, turbulent, non-pulsatile blood flow	• ¹ Fetal distress; • ² trisomy 18, single umbilical artery, arteriovenous fistula

Discussion

Differential diagnosis of an umbilical cord tumor

Nodular bulges of the umbilical cord are rare entities of polymorphous presentation that can be detected prenatally by ultrasound examination. Their differential diagnosis and some of the respective characteristics are listed in Table 1, a more detailed review of which is beyond the this report's scope. The clinical significance common to all of these anomalies is determined by their size, which can potentially cause vascular compromise and affect fetal growth. After birth, umbilical cord clamping should be distal to the lesion to avoid intestinal strangulation. An ultrasound investigation of the lesion is recommended, and referral of the newborn to a pediatric surgery clinic for revision and correction is mandatory, but not an emergency.

Umbilical cord teratoma

Umbilical cord teratoma is a polymorphous, congenital nodular-cystic neoplasm having different cellular and organoid components deriving from more than one germ layer. Its texture varies from soft adipous areas to firm/solid cartilaginous or bony structures characterized by distinct calcification. Teratomas are observed all along the umbilical cord, the origin of which is of normal aspect. Usually considered benign, about one half of all reported cases are associated with other malformations [10].

Hemangioma of the umbilical cord

Hemangioma of the umbilical cord is characterized by well-defined aggregates of closely packed, thin-walled capillary proliferation originating from the umbilical arteries, the

umbilical vein or vitelline capillaries [7]. The etiology is still not clear; it may represent a true neoplasm or it may be a developmental abnormality (hamartoma). Even though it lacks circumscription or encapsulation, it never metastasizes [3], although it has been reported to be associated with additional skin, liver, intra-abdominal and placental hemangiomas. Most often hemangioma is an isolated anomaly, but large lesions have been described in association with polyhydramnios [2], intrauterine growth retardation, elevated maternal serum alpha-fetoprotein [9] and fetal malformations [2]. Some authors have also reported its association with premature delivery and even with fetal death caused by impaired umbilical circulation resulting in nonimmune hydrops fetalis [4], torsion, compression, or stenosis of the umbilical vessels, fetal hemorrhage [7], thrombosis of an umbilical vessel and hematoma of the umbilical cord [11]. We reviewed 37 reports in the literature from 1951 through 2005, in most of which the lesion was referred to as hemangioma, and rarely as angiomyxoma, myangioma or hemangiofibromyxoma because of the myxoid appearance of the commonly associated edematous degeneration of the Wharton's jelly. A possible hereditary predisposition to this vascular anomaly is still under discussion.

Hernia into the cord

Hernia into the cord is a rare, peculiar form of abdominal wall defect differing from the omphalocele by its smaller size and its structure, consisting in a protruding peritoneal segment in the umbilical cord containing no intestinal loop. It develops proximally to the fetal abdominal wall and, typically, the origin of the umbilical cord is of normal size (diameter about 1 cm) and aspect. In the majority of cases it is an isolated finding.

Omphalocele

Omphalocele is the most frequent pathologic entity of those cited above, with an estimated incidence of 1 in 3,000 to 4,000 births. It is a form of abdominal wall defect that is, by definition, between 4 and 12 cm in diameter. It consists of a protrusion of intestinal loops into the base of the umbilical cord, occasionally accompanied by other visceral organs such as the liver, covered by peritoneum without overlying skin. Characteristically, the origin of the umbilical cord is abnormally large. In the majority of cases, omphalocele is associated with other congenital anomalies of the gastrointestinal tract or cardiovascular system, syndromes or chromosomal aberrations.

Cystic lesions

Cystic lesions originate from embryonic vestiges remaining as a consequence of the incomplete obliteration of the urachus and omphalomesenteric duct. They are more often single than multiple findings and are usually isolated, even though their association with other congenital anomalies, usually of the gastrointestinal tract, and other syndromes and sequences has been described. The presence of multiple umbilical cord cysts is associated with an increased risk of miscarriage, aneuploidy and congenital anomalies [5]. Additionally, pseudocysts have also been described, originating as a result of myxoid degeneration of the Wharton's jelly secondary to persistent patent urachus, urachus cysts or abdominal wall defects [1].

Umbilical cord polyp

Umbilical cord polyp results from the persistence of protruding intestinal tissue within the umbilical cord, remnants of the omphalomesenteric duct, which differentiates into proliferating gastrointestinal cells. Communication with the intestinal canal is not always lost. As an isolated finding, it can be a harmless lesion, but its presence should lead to a careful investigation of the possible presence of other omphalomesenteric duct anomalies, such as Meckel's diverticulum and umbilical enteric fistula. The lesion can be exudative and complicated by ulceration and bleeding [6]. There is a report in the literature of an umbilical cord polyp originating from the urachus [8].

Hematoma and aneurysm of the umbilical cord

Hematoma of the umbilical cord is usually an accidental event secondary to cordocentesis done for diagnostic purposes, while spontaneous hematoma remains very rare [11, 12]. Umbilical artery aneurysm is an extremely rare

vascular anomaly associated with poor fetal outcome, comprehending chromosomal aberration, progressive oligohydramnios and fetal death due to acute umbilical venous compression [13].

Conclusion

We report the case of an infant with an umbilical cord tumor that had twice been diagnosed previously: antenatally, by ultrasound, as a teratoma, based on its inhomogeneous character and the absence of an intratumoral flow, and postnatally as a hernia into the cord. The infant was born at term weighing 2,840 g, with a 4-cm diameter nodular bulge in the umbilical cord. The definitive diagnosis was made by histopathology as hemangioma of the umbilical cord.

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